

Mucinous Low-grade Neoplasm of Appendix: A Case Report

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ABSTRACT

Low grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy of the gastrointestinal tract, the most common site being the appendix. Late diagnosed neoplasms may not be differentiated from adnexal masses even by advanced imaging methods and other diagnostic procedures. This might lead to fatal condition. These tumors are often discovered incidentally, either during a survey or at the time of surgery for other causes. We hereby present three cases of LAMNs found incidentally during various surgeries in our centre. We conclude that early histopathological diagnosis is mandatory for timely treatment.

Keywords: Neoplasm; appendix; mucinous.



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INTRODUCTION

Low-Grade Appendiceal Mucinous Neoplasm (LAMN) is a rare epithelial tumor of the appendix, often discovered incidentally during surgery for unrelated conditions. Unlike typical appendiceal tumors, LAMN has the potential to progress to pseudomyxoma peritonei (PMP) if mucinous material spreads beyond the appendix.

Early detection is crucial, as timely surgical intervention can prevent complications and improve patient outcomes.

In this case report, we present three female patients who underwent surgery for different gynecological and surgical indications in our center, during which an incidental diagnosis of LAMN was made following appendectomy. This report highlights the significance of routine appendectomy in surgical procedures, its potential role in early tumor detection, and the importance of histo-pathological evaluation in preventing disease progression.

By documenting these cases, we aim to raise awareness about the incidental discovery of LAMN and discuss its clinical implications.

Case 1: 33 years old female complaining of pain lower abdomen and abdominal distension since 3 months. Physical examination showed firm, mobile abdominal mass with slight tenderness. There was no significant past history of abdominal surgery. Ultrasonography of abdomen and pelvis showed a blind loop-like structure

within the right iliac fossa. A hypoechoic lesion was seen within the appendix. CT scan showed an obstructive non-enhancing lesion at the base of the caecum without invading the caecum. The surgery was done. Specimen of terminal ileum, caecum, ascending colon and part of transverse colon and appendix were sent for histopathological examination.

Case 2: A 54years old female presented with the complaints of pain abdomen and occasional fresh, blood-stained discharge per vaginum. On physical examination, there was a well-defined, non-tender hard mass per abdomen. CT scan showed a large complex right adnexal cystic mass(9.2x11cm) and a non-enhancing cystic lesion. Thus, clinical presentation and radiological findings initially pointed toward a gynecological issue, confirmed by imaging as a large complex right adnexal mass. The presence of a dermoid cyst led to surgical intervention, including hysterectomy and salpingectomy.

However, the unexpected finding of an enlarged appendix during surgery raised suspicion of an appendiceal neoplasm. This prompted a right hemicolectomy with appendicectomy and colonic margin resection, likely to ensure complete removal in case of malignancy.

Case 3: A 72-year-old female complained of fresh per vaginum bleeding on and off for 2 months. Physical examination showed adnexal mass. Radiological findings showed well defined,

hypoechoic solid mass within the endometrial cavity and panhysterectomy was planned for fibroid uterus. On surgical intervention, submucosal leiomyoma was observed. Additionally, appendectomy was done with the clinical suspicion of appendicular neoplasm.

Gross: In all three cases, appendix showed grayish-brown discoloration, grossly dilated, smooth and congested outer surface. The cut surface showed chalky white mucinous and granular material in the lumen.



Figure 1(a): Outer surface of appendix showing grossly dilated, grayish brown discoloration and smooth congested surface.



Figure 1(b): Cut surface of appendix showing mucinous chalky white and granular material in the lumen.

Microscopy: Tall columnar mucinous lining epithelium pushing into underlying tissue with villous, serrated and underlying architecture was observed. The epithelial cells and glands showed single to 2-3 layers of epithelial stratification with elongated, columnar, bland to hyper-chromatic nuclei and abundant mucin vacuoles in the cytoplasm. The neoplastic epithelium cells were resting on fibrous stroma. Absence of lymphoid follicles and extensive areas of calcification were observed in the latter two cases. Extracellular mucin comprised of > 50% of the lesion and extended upto periappendicular area forming a well circumscribed lesion. The serosal layer pools of mucin contained inflammatory cells mainly lymphocytes and plasma cells with paucity of

epithelial cells. The final histopathological diagnosis of LAMN (pT3 Nx Mx) was considered in all three cases.

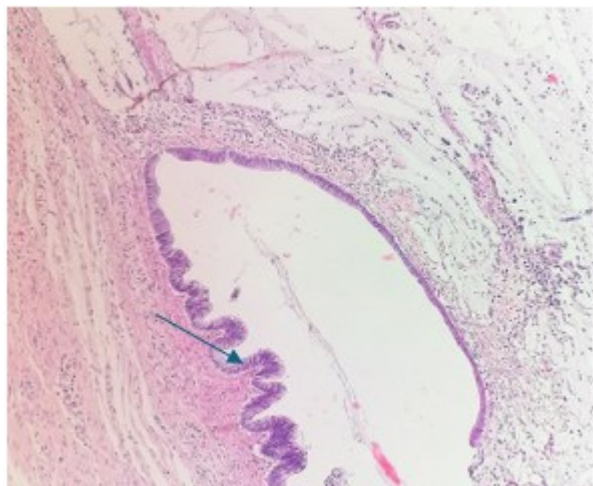


Figure:2 The lining epithelium shows villous epithelium. (Arrow)

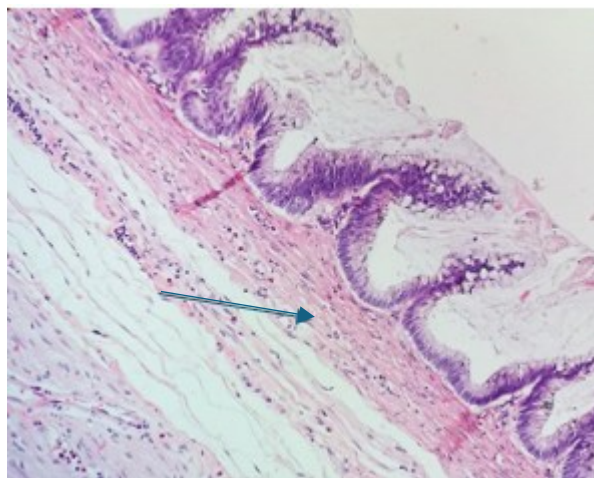


Figure: 3 The neoplastic epithelium rests on fibrous stroma (Arrow). There is absence of lymphoid follicles.

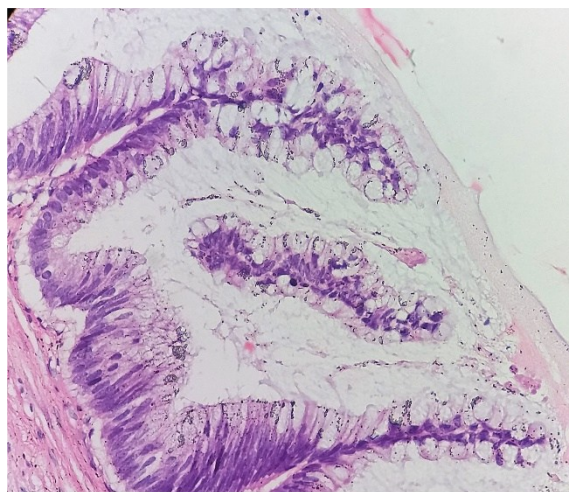


Figure 4: Neoplastic cells have columnar to cuboidal cells with bland to hyperchromatic nuclei and mucin vacuoles in the cytoplasm.

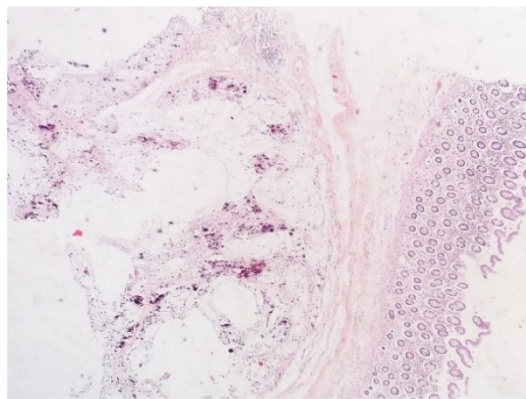


Figure 5: Extensive areas of calcification

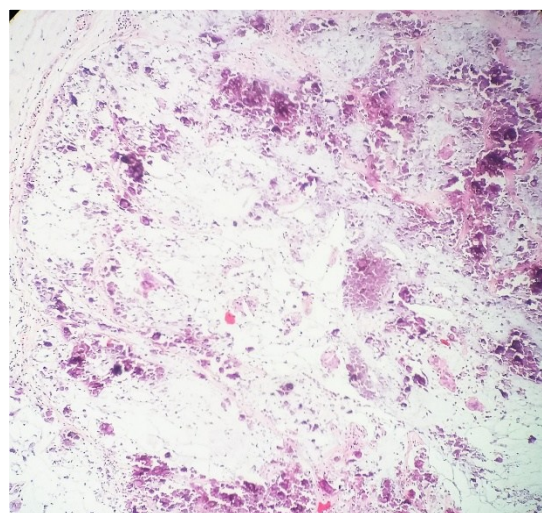


Figure:6 Extracellular mucin comprises >50% of the lesion and is extending upto periappendiceal area forming a well-circumscribed lesion and pushing the caecal wall.

DISCUSSION

LAMN was first described by Rokitansky in 1842.[1] It is a rare entity found in 0.3% of appendiceal specimens.[2] As per the similar study done by Dixit et al., LAMNs have a predilection for females above the age of 50 and merit inclusion in the differential diagnosis of acute or chronic right lower quadrant abdominal pain. In the early stages of the disease, symptoms may be presented as right lower quadrant pain, which can be attributed to mucin-induced distension of the appendix.[3] In our index case also, there was female predilection, and all the cases had abdominal pain while two of the cases also presented with vaginal bleeding. During the surgical management for the suspected gynaecological pathology, appendicectomy were done additionally for the suspected appendiceal swelling in the right adnexa in all three cases in our study.

In a study done by Emre et al., mean age of a total of 13 cases including six women (46%) and seven men (54%) was 52.4+/-21.6 years (Range :25 to 83 years).[4] In our present study, the range of the patients were from 33 to 72 years of age.

Microscopically, classical LAMNs show replacement of the normal appendiceal mucosa by filiform or villous hypermucinous epithelium with cytoplasmic mucin vacuoles and basally placed nuclei with minimal to mild atypia. An accurate histopathological analysis of the entire appendiceal specimen is critical to ensure an accurate diagnosis. Similar histomorphology features were seen in the present case.[5] Specifically, the identification of epithelial invasion within the appendiceal wall is pivotal for distinguishing adenocarcinomas from other mucinous lesions. The presence of this key histological indicator can be challenging to ascertain as overt invasion may not be evident and may instead present as fibrotic attenuation of the mural layers, with or without mucin penetration. [6]

Appendiceal mucinous lesions are histologically categorized as non-neoplastic or neoplastic epithelial lesions. The neoplastic forms can be further subdivided into serrated polyps, hyperplastic polyps, appendiceal mucinous neoplasm (AMN) and mucinous adenocarcinomas. World Health Organization (WHO) classification of Digestive System tumors, (5th Edition, 2019) classified the AMN into two groups based on the degree of cytological atypia, Low grade appendiceal mucinous neoplasm (LAMN) and high grade appendiceal mucinous neoplasm (HAMN). [5]

The complications of LAMN include intussusception, volvulus, small bowel or ureteral obstruction. [5] There is always the risk of rupture, either spontaneous or accidental, with consequent development of pseudomyxoma peritonei. [6]

LAMN many times mimic ovarian carcinoma as they both present with pelvic/abdominal pain.[7]

Although a mucocoele or LAMN may be diagnosed pre-operatively with abdominal USG, CT or MRI, diagnosis is generally made intraoperatively due to macroscopic appearance and a final diagnosis can only be reached after histopathological examination of the specimen. [4]

Metastatic LAMN is an aggressive disease and requires cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) whereas low-grade mucinous tumors are surgically curable. [5]

It is therefore very important to classify LAMN. Mucin extending into the muscularis propria should be classified as pTis (LAMN) as long as it does not extend into the mesoappendix or serosa. LAMNs showing either acellular mucin or mucinous epithelium involvement to subserosa or mesoappendix are classified as pT3. Perforation of the appendix by LAMN is associated with a high risk of peritoneal dissemination. Given the risk of disseminated peritoneal mucinous disease, pT4 includes assessing both acellular mucin and the

mucinous epithelium and is designated as T4a (penetration of the serosa) and T4b. [8]

Early diagnosis and optimal intervention for treatment are thus necessary in leading to a good prognosis. Since histopathology is the mainstay in the diagnosis, pathologists should be very vigilant during their examination.

CONCLUSIONS

All of the cases were discovered as incidental findings at appendicectomy, during laparotomy for another indication, or during histological examination of an operative specimen. The main purpose of this study was to describe the clinical and histomorphological features of low grade appendiceal mucinous neoplasms. This study highlights the fact that patients can benefit from early histopathological diagnosis of appendiceal mucinous neoplasms which warrants immediate intervention measures.

CONFLICT OF INTEREST

None.

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None.

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